Cronkhite Canada Syndrome with Early Colorectal Carcinoma in a Patient

Abstract:
C Mason 1, C Quinlan 2, M O’Donovan 1, J Hyland 2, C Fallon 1
1 Midlands Regional Hospital, Mullingar, Co Westmeath
2 St Vincents University Hospital, Elm Park, Dublin 4

We relate the first case in the Republic of Ireland of Cronkhite Canada Syndrome (CCS). The patient presented with weight loss, alopecia, nail dystrophy, taste disturbance and classic radiologic and endoscopic features of CCS. She continued to dramatically lose weight and early repeat colonoscopy showed the interim development of an invasive sigmoid adenocarcinoma.

Case Report
A 66 year old white Irish female was referred complaining of loss of appetite, taste disturbance, alopecia and dystrophic nails. She had an unremarkable medical history. She was an ex-smoker of 30 years and was on no medications. Clinical examination revealed a body mass index (BMI) of 19kg/m2 and evidence of dystrophic nails and alopecia. Blood tests were entirely normal including full blood count, thyroid, renal, liver and bone testing, erythrocyte sedimentation rate (ESR), serum calcium and serum ferritin. Endoscopy demonstrated a grossly abnormal stomach with a mass of polypoidal material prominent in the antrum along with multiple duodenal polyps. Helicobacter pylori test was negative. Biopsy from the gastric material demonstrated an inflammatory fibroid polyp while the duodenal polyps showed marked flattening of villi and acute inflammation but a normal intra-epithelial lymphocyte count.

Colonoscopy showed polyposis of the entire colon. Serial biopsies showed inflammatory infiltrates with neutrophils, prominent eosinophils and submucosal oedema. Barium swallow and small bowel follow through confirmed grossly abnormal mucosa of the stomach along with generalized thickening of the mucosal folds of the duodenum and jejunum and filling defects in the ileum suggestive of small bowel polyposis. She was commenced on folic acid, multivitamins, dietary supplements and lanzoprazole. Zinc supplements were prescribed for the taste disturbance. On review shortly afterwards, hair and nail growth had returned, but dysgeusia and significant weight loss continued. In view of the ongoing weight loss, a repeat colonoscopy was performed less than 2 months after the previous examination. On this occasion, along with the colonic polyposis, there was now a suspicious irregular polyp at 25cms. Histology confirmed an invasive moderately differentiated adenocarcinoma. She subsequently underwent surgical resection for a T4N2M0 tumour and was referred for adjuvant chemotherapy.

Discussion
Cronkhite-Canada syndrome (CCS)1, is a rare, acquired, nonfamilial syndrome of diffuse gastrointestinal polyposis, atrophic nail change, alopecia, taste disturbance, cutaneous hyperpigmentation, diarrhoea, protein-losing enteropathy and malnutrition. We report a 66 year old white Irish female presenting with the typical clinical, endoscopic, radiologic and histologic features of CCS. This is the first reported case from the Republic of Ireland. There are over 400 cases in the literature, close to 75% of which originate from Japan2,3. There have been very few reported cases in the British Isles4,5. The one previous reported case on the Irish mainland was in an elderly Japanese male who died shortly after presentation5. The age of onset of CCS is typically during the 6th or 7th decades. The cause remains unknown although there have been associations with physical and psychological stress2,3, autoimmune disease, Helicobacter pylori infection and pregnancy.

Gastrointestinal manifestations include recurrent abdominal pain, chronic or episodic diarrhoea and often profound weight loss due to altered digestive, motive and secretory functions of the bowel and associated bacterial overgrowth. Polyposis principally in the colon but also in the stomach and small intestine is typical. The ectodermal manifestations of alopecia, nail dystrophy, and hyperpigmentation are characteristic. Taste disturbance (dysgeusia) is often prominent as it was in our patient and sometimes responds to zinc7. While CCS was initially thought to be a benign disease, it is now recognised as a pre-malignant state with malignancy rates estimated at up to 20%. The malignancy risk appears to be confined to the gastrointestinal tract. Due to the rarity of the disorder, there are no

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clear guidelines as to how often endoscopic surveillance should be performed. Patients usually have progressive symptoms but may have spontaneous remissions, making it difficult to attribute any improvement to a specific therapy. Prognosis is usually poor with a reported 5-year mortality rate of 55% with mortality due to malignancy, sepsis and gastrointestinal blood loss. Current treatment strategies have not been proven to be effective. There are interesting anecdotal cases of remission with aggressive and prolonged nutritional supportive care, anti-Helicobacter treatment and especially with corticosteroid use9,10.

Correspondence: C Mason
BOX 135, Department of Medicine for the Elderly, Addenbrookes Hospital, Hills Rd, Cambridge, CB2 0QQ
Email: colin.mason1@addenbrookes.nhs.uk

References